

RESEARCH PAPER

Nutrition education tools used in phenylketonuria: clinician, parent and patient perspectives from three international surveys

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Abstract**Background:** Three international surveys were developed aiming to identify the current nutrition educational tools used in the management of phenylketonuria (PKU) and the perceived effectiveness of these tools by clinicians, parents and patients.**Methods:** The first two surveys were distributed through the Metabolic Dietitians ListServe (pno-metabl@listserv.cc.emory.edu), and the third survey was distributed by international clinics and the National PKU Alliance website (www.npkua.org). A total of 888 responses (S1, $n = 88$; S2, $n = 81$; S3, $n = 719$) were collected from all three surveys. The surveys represent participants from 17 countries, in Europe; North America (USA and Canada); Mexico; Argentina; Turkey; Australia; and Africa (Tunisia).**Results:** A consistent decline in 'parents as role models' as an educational tool was observed starting at age 10 years. Patients responded they feel their families are the most effective form of education, whereas handouts were selected as the least effective educational tool by patients. Parents responded they feel the most effective educational tool is one-on-one counselling. Patients and parents show a desirable trend in wanting to attend group clinic, even in centres where this type of educational tool is not offered.**Conclusions:** There was a discrepancy between clinicians and patient views regarding the perceived effectiveness of the nutrition education tools. Future research is needed surrounding the impact nutrition education may have on improved dietary compliance in patients with PKU.**Introduction**

The long-term dietary management of Phenylketonuria (PKU) is a complex and well documented challenge for many patients to adhere to as a life-long treatment (MacDonald, 2000; Blau *et al.*, 2010; MacDonald *et al.*, 2010). Outcomes are related to the level of adherence and compliance to the treatment. Educational tools have been developed and are currently used in the treatment

and management of the diet. Current nutrition education tools include (but are not limited to) one-on-one counselling, handouts/printed material, support groups, cooking classes, seminars, group clinics, parents as role models, and computer-based software. Studies have indicated that familial support, practical nutrition education and active involvement and/or intervention with parent and patients can improve dietary compliance and quality of life in chronic conditions (Hanson *et al.*, 1995;

Cochran & Conn, 2008; Trahms *et al.*, 2010). Various quality of life aspects, including familial, socio-economic, cultural and emotional, play a role in evaluating the efficacy of nutrition education tools currently used by clinicians. Similar factors have been found in patients with arthritis, cystic fibrosis, galactosemia, coeliac disease and diabetes, and are often described as a burden of a chronic condition (Bosch *et al.*, 2004; Llorente *et al.*, 2008; Wagner *et al.*, 2008; Kratz *et al.*, 2009; Hansen *et al.*, 2012). As described by Walter *et al.* (2002), dietary compliance in PKU decreases with age, further impacting upon quality of life. Currently, there are no studies in PKU describing the impact nutrition knowledge may have on the outcome of dietary compliance.

The present study aimed to identify the educational tools currently used by clinicians and their perceived effectiveness by professionals, parents and patients through results obtained from three international surveys. Additionally, it study intended to draw attention to the perceived efficacy of current nutrition education tools, rather than provide recommendations for PKU nutrition education and improved compliance.

Materials and methods

Three surveys were developed to identify the current nutrition education tools used by clinicians and the perceived effectiveness of these tools by professionals, parents and patients. Survey responses were anonymous and each survey had an independent set of respondents. Clinicians may have responded to either survey 1 or survey 2. Parents and patients responded solely to survey 3. Incomplete surveys were discarded from analysis.

The first survey (S1), International Survey of Educational Tools Used in PKU Dietary Compliance ($n = 88$), asked clinicians to define the term dietary compliance and to identify the types of current nutrition education tools currently used in their clinics that specialise in treating patients with PKU from 3 to 21 years of age. The results from S1 were assembled to create the second survey (S2), International Survey of Perceived Effectiveness of Educational Tools Used in PKU Dietary Compliance ($n = 81$), in which clinicians were asked to evaluate the perceived efficacy of the nutrition education tools identified in S1.

In S1 and S2, the questions were constructed as: yes/no, multiple choice, and many questions had to option for individual typed responses. In several questions, the respondents had the opportunity to select, in order of decreasing importance, as many answers as they felt were applicable.

The third and final survey (S3), Parents and Patients Perception of the Effectiveness of Nutrition Education

Tools Used in PKU Dietary Compliance, gathered information and perceptions from patients and parents ($n = 719$) regarding the effectiveness of nutrition education tools and the impact that these tools may or may not have on dietary compliance.

All surveys were constructed on the secure, web-based, SurveyMonkey (<http://www.surveymonkey.com/>) questionnaire platform and distributed via a secure website link. The results from S1 and S2 represent participants from clinics in 12 countries in Europe; North America (USA and Canada); Mexico (S1 only); Argentina; Turkey (S2 only); and Australia. Clinicians were contacted via e-mail, ListServ and websites and asked to complete the first two surveys (S1 and S2). The patients and parents were contacted by the clinics and parent associations, asked to complete the survey, and given a secure web link (S3) to complete the survey. In the USA, S3 was also distributed through the National PKU Alliance (NPKUA, <http://www.npkua.org>) website. S1 and S2 were provided in the English language only to clinicians. S3 was provided in French, Polish, English and Portuguese for patients and parents. The results from S3 represent participants from five countries in Europe; Norway; Tunisia; Australia; and North America (USA and Canada).

Statistical analysis was completed using *t*-tests in EXCEL (Microsoft Corp., Redmond, WA, USA) and using R software (R Project for Statistical Computing, Vienna, Austria) for linear and multiple regression analysis of the data. R software comprises an integrated suite of software facilities for data manipulation, statistical computing, calculation and graphical display. $P < 0.05$ was considered statistically significant.

Results

Surveys 1 and 2: International survey of educational tools used in PKU dietary compliance by clinicians

Definition of compliance

Eighty-eight clinicians participated in S1 and 81 clinicians participated in S2. The data from S1 and S2 indicate that 94.7% of clinicians agreed that dietary compliance could be defined using the criteria: Phe levels in treatment range (92.1%), regular monitoring of Phe levels (86.5%) and completing prescribed metabolic formula (80.9%). The combination of the top three choices makes the definition of compliance statistically significant over all other choices ($P = 0.027$). Other terms encompassing the definition of dietary compliance included clinic attendance (73.0%), consuming the proper amount of Phe per day from food (64.0%) and keeping diet records (32.6%).

Current educational tools

S1 concluded that 80.9% of surveyed clinicians currently offer PKU nutrition education in their clinics in addition to regular counselling. Clinicians were asked to select the types of nutrition education tools currently used within their respective clinics. Responses included one-on-one counselling, parents as role models, printed material/handouts, cooking classes/demonstrations, seminars, parent group clinic, patient group clinic, and peer-to-peer counselling. Clinicians responded that nutrition education is primarily performed by one-on-one counselling (97.8%) and by printed materials/handouts (84.4%). Parent seminars (28.9%), parent group clinic (24.4%) and peer-to-peer counselling (17.8%) were utilised significantly less than when a combination of one-on-one counselling and printed material/handouts were used as an educational tool ($P = 0.034$).

Education and compliance

Clinicians were asked whether they felt there was a correlation between the type of nutrition education tool used and dietary compliance in their patients. The majority of clinicians (86.1%) responded 'Yes', the type of tool used does impact dietary compliance. Of the clinicians in agreement, 88% responded that poor parental involvement is the primary reason why nutrition education tools do not correlate to increased dietary compliance in patients.

Introduction of nutrition education

The responses varied when asked at what age clinicians begin nutrition education for their patients with PKU. Of all the clinicians that responded to S1 and S2, 54.4% begin educating their patients about PKU nutrition between the ages of 3–5 years, followed by ages 6–9 years (35.6%); some begin education as late as 10–12 years (7.8%). We found that nutrition education in all responding countries was started before age 12 years.

Educational tools: types and perceived efficacy

In S1, clinicians were asked 'Which educational tool(s) are currently used in your patient population?'. We observed a consistent decline in 'parents as role models' as an educational tool starting at age 10 years (Fig. 1). Across all age groups, clinicians selected group clinics as the *least* utilised nutrition education tool.

Building off of the responses in S1, clinicians in S2 were asked 'Which educational tool do you perceive to be the most effective in dietary compliance in patients with PKU?'. One-on-one counselling was perceived to be the most effective tool of all of the choices, used across all age groups ($P = 0.045$), followed by group clinic until age 15 years. By age 16 years, clinicians responded cooking classes/demonstrations take the place of group clinic as being perceived as the second most effective educational tool. Clinicians in S2 perceived handouts to be the least effective tool across all age groups (Fig. 2).

Group clinic

Ten of the 17 countries responding in S2 currently do not offer group clinic as an educational tool. Although this number is not statistically significant ($P = 0.059$), it is noteworthy. Of the seven countries that offer group clinic, most of these clinics offer it only 13% of the time. Clinicians were asked 'Do you believe group clinic is an effective nutrition education tool?'. Of those asked, 84.5% perceive group clinic as an effective nutrition education tool ($P = 0.033$). Conversely, 15.5% perceive group clinic as an ineffective tool. Lack of patient population, inadequate facilities and no experience with facilitating group clinics were the open-ended reasons reported as to why group clinic is perceived as an ineffective educational tool.

Barriers to compliance

Clinicians were asked 'In your opinion, please select all factors that may contribute to dietary noncompliance'.

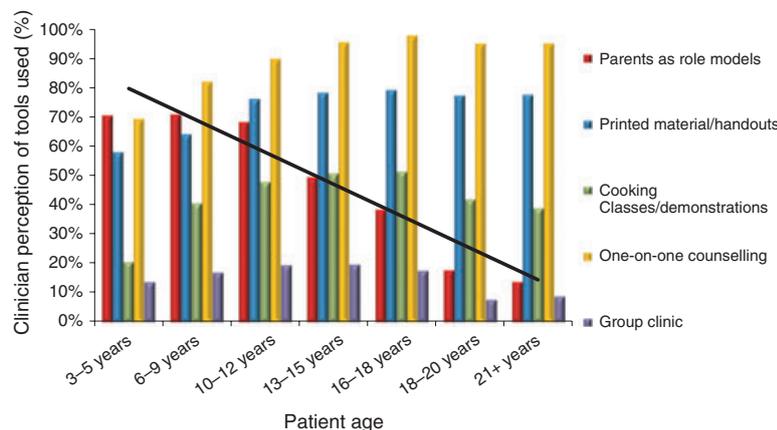


Figure 1 Clinician perception of educational tools used by age (S1).

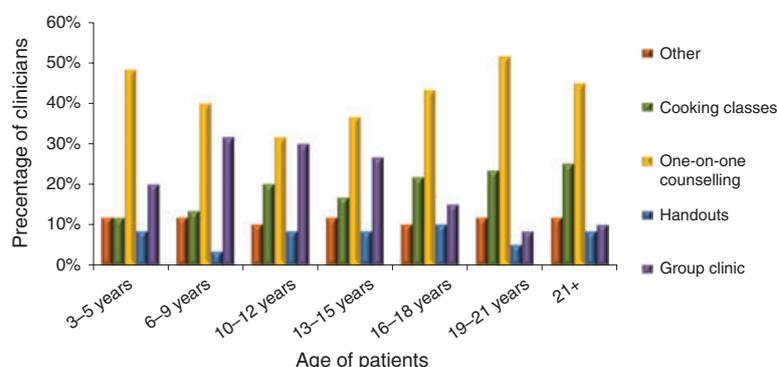


Figure 2 Perceived effectiveness of educational tools by clinicians (S2).

Embarrassment and/or frustration around the diet (90%), poor family cohesion (83.3%) and difficulty in food preparation (78.9%) are the top three barriers to compliance reported by clinicians (Table 1).

There are several noteworthy points specific to country and geography: Mexico was the only country in which clinicians did not select 'embarrassment' as a barrier to compliance; clinicians in Portugal, Denmark, Sweden, Ireland, UK and Australia did not select 'lack of insurance coverage', and 'poor family cohesion' was not selected as a barrier to compliance by clinicians in Ireland.

Utilising the answers from S1 (Table 1), the authors asked clinicians in S2 'Which of the following factors do you perceive as the greatest barrier to compliance in your patients?'. Inconvenience of diet was perceived as the most prevalent barrier (52%), followed by poor family cohesion, lack of insurance and the limitations of a phenylalanine restricted diet (45%) (Fig. 3). It is noted that clinician responses in S1 did not include inconvenience of diet or limitations of a phenylalanine restricted diet as potential barriers that may contribute to noncompliance.

Table 1 Potential contributing factors to noncompliance (S1)*

| | Response percent (%) |
|--|----------------------|
| Embarrassment and/or frustration around the diet when participating in social activities | 90.0 |
| Poor family cohesion | 83.3 |
| Difficulty in food preparation | 78.9 |
| Socio-economic status | 76.7 |
| Lack of insurance coverage for medical formula, medication and low-protein foods | 64.4 |
| Role models compromise compliance (teachers, counsellors, babysitters, etc.) | 61.1 |
| Language and cultural barriers | 55.6 |
| Availability of dietary treatment | 31.1 |

*These results are any factors clinicians perceive be a contributor to noncompliance (multiple answers allowed).

Computer-based tools

The authors asked clinicians whether they considered computer-based applications as a nutrition education tool would improve long-term dietary compliance. Some 94.8% agreed that it would be effective for patients ranging from 5 to 18 years, whereas 86% agreed that it would be effective for ages 19–30 years, and still half (54.4%) considered a computer application would be effective for patients aged >31 years. Clinicians responded in agreement (96.6%) that, if a computer-based nutrition education tool were available, they would use it.

Survey 3: Parents and patients perception of the effectiveness of nutrition education tools used in PKU dietary compliance

Subsequent to the results from S1 and S2, the authors developed a third survey focusing on patients, parents and caregivers. S3 gathered information regarding the perceived effectiveness of nutrition education tools used in the treatment of PKU.

Seven hundred and nineteen responses were obtained from 12 countries. Four hundred and forty-seven responses from parents and 272 responses from patients were analysed. Clinicians did not participate in S3. For both parent and patient surveys, 73% were female. The mean age of patients completing the survey was 19.9 years (range 6–54 years).

Definition of compliance

Statistically significant is the agreement by parents and patients ($P = 0.030$, $P = 0.039$, respectively) with the definition of dietary compliance from S1 and S2 (Phe levels within treatment range, consuming prescribed metabolic formula and regular monitoring of Phe levels). The 98% of parents and 96% of patients agreed nutrition education is important for dietary compliance and in their overall health ($P = 0.040$).

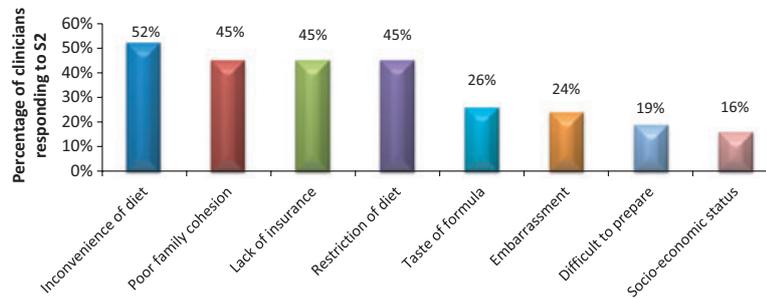


Figure 3 Clinician's perceived greatest barriers to compliance (S2).

Educational tools: types and perceived efficacy

Handouts, one-on-one counselling and cooking classes were selected as the most common types of nutrition education both parents and patients have received from clinicians. This is in alignment with the results from S1 and S2, where one-on-one counselling and handouts were selected as the preferred tools by clinicians. Parents in all countries and languages responded that one-on-one counselling is perceived to be the most effective tool. Patients responded that they consider their families are the most effective educational tool (58.6%). Seventy-eight percent of patients selected handouts as the least effective tool currently used, which is in direct contradiction to the results obtained from S1 and S2.

Group clinic

Group clinic in S1 and S2 was determined by clinicians to be an important educational tool, although few clinicians offer this as an educational option (Fig. 2). Of interest were the different responses received from Polish parents and patients versus all other responses. Polish parents and patients responded that group clinic is offered by 68% of their respective clinics. Excluding the Polish responses, group clinic is only offered, on average, by 39% of all other international clinics. Parents were asked whether their child had attended a group clinic and 67% of Polish patients had attended, whereas only 51% of all other patients had ever attended a group clinic. Eighty-eight percent of all responding patients and 91% of all responding parents said they would attend a group clinic if it was offered. Both patients and parents show a desirable trend in wanting to attend a group clinic.

Barriers to compliance

Results were mixed when patients were asked what they feel is the primary barrier to being compliant with their diet. In S1 and S2, 'embarrassment' was selected by clinicians as a barrier to compliance, although Polish and English speaking patients noted 'being different from friends' and 'curiosity of new foods' as top barriers to dietary compliance. French and Portuguese patients specifically noted 'taste of formula' as the top barrier to

dietary compliance. English speaking patients also noted a 'lack of family support' and 'lack of access to low phenylalanine foods' as barriers to compliance, which aligns with the clinician perceptions found in S1 and S2.

Computer-based tools

The use and frequency of computer-based application was asked of both parents and patients in S3. Polish parents and patients had the greatest percentage of computer-based application use, followed by Portuguese, English and French. Computer-based applications are used by the greatest percentage of Polish parents and patients (82.1% and 78.8%, respectively). The 60.8% of Portuguese parents and 39.3% of patients report using computer applications, whereas only 32.4% of English speaking patients and 16.5% of French speaking patients report using a computer application as an educational tool to help manage their PKU diet. A trend was identified with the use of computer-based tools and patient reports of cheating on the diet. It appears that a greater reported use of computer technology positively correlates with a lower admission of cheating on the diet.

Discussion

The results obtained in the present study show the majority of international clinics offer PKU nutrition education. Although the definition of compliance was almost consensual, there was a discrepancy between clinicians and patient views regarding the type of nutrition education tools used and the perceived effectiveness of these tools.

The gaps and challenges presented in these surveys highlight the need to develop educational tools that are individualised and appropriate for age and level of learning. Currently, few alternative educational resources are available to help fill these educational gaps. As clinicians, a modification to the current nutrition education tools used in conjunction with one-on-one counselling may be the best approach to meet the needs of the patient throughout his/her lifecycle.

The perceived influence of family involvement in the adherence to the PKU diet was significant because

clinicians (89.7%) selected family as 'greatly influencing compliance' ($P = 0.023$). The results of the present study echo similarities from studies that show increasing the level of responsibility beginning in the early school age years will help facilitate the patient to understand and manage his/her condition.

In addition, early nutrition education gives the patient a greater sense of control with his/her diet and allows parents to transfer the care of responsibility (Demirkol *et al.*, 2011; van Spronsen *et al.*, 2011). Beginning nutrition education at an early age is also supported by the research carried out by Olsson *et al.* (2007), who noted that parents bear the main responsibility for adherence throughout childhood and promote adherence in their children affected by PKU. Our results show that one-on-one counselling and handouts are well-received tools by the parents. S1 and S2 confirm the data presented by MacDonald *et al.* (2010); where open discussions and explanations, together with sufficient time to answer questions, which are aspects of one-on-one counselling, are invaluable to dietary compliance. Written educational resources are commonly used in addition to one-on-one counselling, although they have been shown to be less effective than other educational interventions (Roter *et al.*, 1998; Durham-Shearer *et al.*, 2008). Responses from clinicians (S1 and S2) and parent and patient surveys (S3) show that handouts were selected as the *least* effective tool, despite being selected as one of the most common educational approaches used by clinicians (S1 and S2; Fig. 1). Interestingly, in the study performed by Jeffries & Vibhuti (2011), it was noted that, when provided with different educational tools, clinicians prefer to learn from pocket cards and seminars (i.e. tools that are simple and readily accessible). Clinicians may need to examine why they are utilising handouts with patients. A possible reason may be because it is a familiar and typically utilised during daily clinical routines. The challenge to clinicians is to create and provide an effective handout, in conjunction with additional educational tools (i.e. one-on-one counselling), which interrelate to time and cost.

The surveys also support the data presented by MacDonald *et al.* (2010), where barriers to compliance were found to include inconvenience, poor family support, and conflicting family beliefs. Family dynamics, specifically poor family cohesion, may need to be taken into account with regard to the type of nutrition education tool used for a specific patient. The family's perceptions, values and responses are inextricably involved in the management of health, the selection of treatment, and the effectiveness of the educational programme. Critical to the success of the patient is the education that the parents receive during their child's early years because our surveys showed

patients are learning the most from their parents during these formative years.

Insurance availability and coverage can influence product accessibility. A lack of accessibility can influence dietary compliance, which directly affects patient outcome. For example, in the USA, only 28 states have implemented legislation for mandating reimbursement for medical foods and formula (Huntington & Buist, 2009). This may account for the reason why 21.5% of English speaking parents in the USA ($n = 265$) selected 'lack of insurance coverage' as a barrier to compliance. Conversely, in countries such as Portugal and France, the socialised system of medicine coverage includes all types of formulas and food; consequently, a 'lack of insurance' was not selected as a barrier to compliance by parents. An interesting outlier to note is that 49.3% of Polish parents responded a 'lack of insurance' as a barrier to compliance despite their socialised healthcare system.

Our survey results continue to confirm the previous findings from numerous studies regarding factors relating to dietary compliance such as: difficulty in food preparation (Bilginsoy *et al.*, 2005; Demirkol *et al.*, 2011), educational level of parents and caregivers (MacDonald *et al.*, 2010), poor family cohesion (Ipsiroglu *et al.*, 2005; Olsson *et al.*, 2007; van Spronsen *et al.*, 2011) and being different from friends (Nevens, 2005).

Group clinic is perceived to be an effective educational tool based upon responses collected in these surveys, although this term requires more clarification regarding what constitutes a 'group'. Our findings support previous data showing that, as a result of the high level of patient participation required, conflict with work and parent schedules, distance to clinic, and family communication (Durham-Shearer *et al.*, 2008), it is likely group clinic is only feasible for a minority of the patient population, despite the majority of patients and parents expressing an interest in attending group clinics.

In summary, a population of international clinicians, patients and parents were surveyed to determine the current educational tools and the perceived effectiveness of these tools in the management of PKU. It is apparent from the findings that noncompliance is a secondary result to long-term management for a variety of reasons. Present teaching approaches are not well over-lapped with the needs of our patients and parents. For nutrition education tools to be effective, it is possible clinicians need to look outside the current paradigm and reassess their approach.

There are limitations to the data as a result of the survey research method used for data collection. The population pool was diverse without a controlled number of responses from each country. Individuals were erroneously excluded as a result of human error, leading to an

undercount in our survey responses from various countries. To account for this error in our statistics, the percentage of a country's given answer out of the total of completed survey responses was used. The varying number of responses still may have affected the results. Sampling errors encountered include a failure to complete entire survey by respondents, content/reporting errors, and interpretation/translational discrepancies. Access to clinic and/or computers may have also excluded certain demographics. The level of parental participation or supervision of patients completing the survey is unknown.

These surveys were designed and executed with the intent of data collection only. The authors conducted these surveys as a pilot project that necessitates further research to provide guidance to clinicians. Future studies comparing same-age patient populations in international clinics, who have been exposed to various educational tools, are warranted in determining how specific educational tools impact compliance and long-term outcomes.

We did not use hypotheses as a base for the statistical analysis; however, our *P*-values indicate a significant difference and correlation between response number values. Recognising that the results obtained from surveys of this kind are not typically analysed, the *P*-values are derived from the appearance of the number of responses to a particular answer. *P*-values show a significant difference and/or correlation between response number values (e.g. 45 responses to answer A, and 15 responses to answer B were compared for statistical importance). Hypotheses were not formulated because we wanted to objectively collect survey data. The authors recognise that this is a limitation of the survey when analysing the results.

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Terminology

The term 'respondents' refers to clinicians throughout the paper, whereas, patients and parents are identified as such. The term 'parents' encompasses any person who is caring or has cared for a patient with PKU. A parent

could include mother, father, grandparents, aunt, uncle, cousin, etc., and is not specific to the biological mother and father of the patient. 'English' refers to any parent or patient who speaks or writes the English language regardless of residence.

Conflicts of interest, sources of funding and authorship

The authors declare that there are no conflicts of interest. The authors wish to acknowledge that there was no source of funding or financial support for these surveys. LEB conceived and designed the study, analysed and interpreted the data, and acquired, drafted and revised the manuscript. JRH assisted in the design and translation of the surveys, analysed and interpreted the data, and drafted and revised the manuscript. JCR assisted in the design of the study, interpretation of data, and the drafting of the manuscript, and also revised it for important intellectual content. MFA assisted in the design of the study, translation of the surveys and revised it for important intellectual content. FF assisted in the translation of the surveys and revised the paper for important intellectual content. RML assisted in the translation of the surveys and revised the paper for important intellectual content. MG assisted in the translation of the surveys and revised the paper for important intellectual content. All authors critically reviewed the manuscript and approved the final version submitted for publication.

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